PART XXVII The Nervous System

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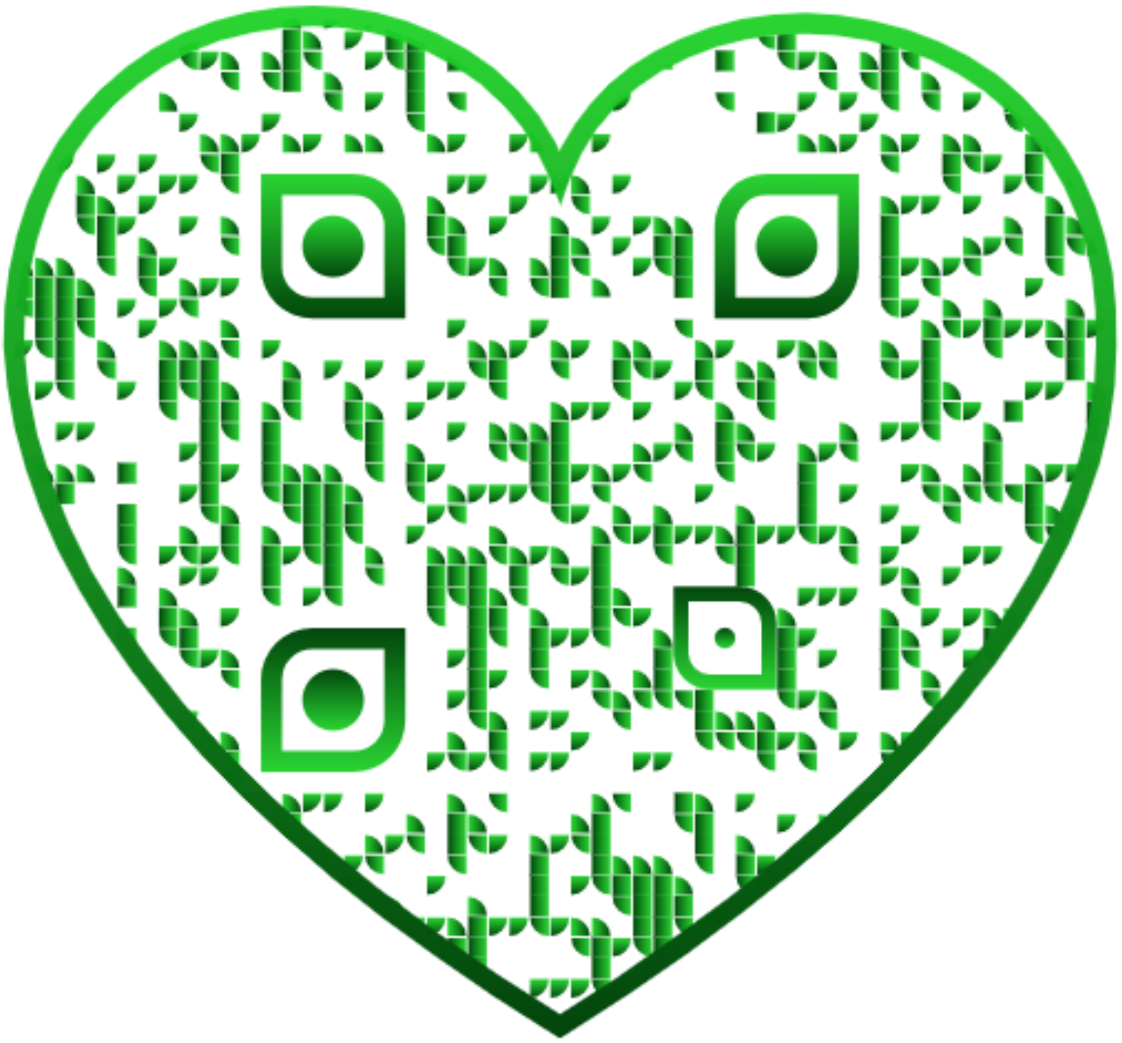
1. The skull tends to assume a square or boxlike shape in
   A. Kleinfelter syndrome
   B. neurofibromatosis
   C. storage disorder
   D. chronic subdural hemorrhages
   E. hydrocephalus

2. Smell can be tested reliably at
   A. 32nd wk of gestation
   B. birth
   C. three months of age
   D. six months of age
   E. nine months of age

3. Contraindications to performing a lumbar puncture include the following **EXCEPT**
   A. suspected mass lesion of the brain
   B. suspected mass lesion of the spinal cord
   C. symptoms and signs of impending cerebral herniation
   D. skin infection at the site of the lumbar puncture
   E. thrombocytopenia with a platelet count $<50 \times 10^9/L$

4. An elevated polymorphonuclear (PMN) cells count in cerebrospinal fluid (CSF) suggests
   A. tuberculous meningitis
   B. early phase of aseptic meningitis
   C. fungal meningitis
   D. demyelinating diseases
   E. brain or spinal cord tumor

5. Xanthochromia of cerebrospinal fluid (CSF) suggests the following **EXCEPT**
   A. bloody tap
   B. subarachnoid hemorrhage
   C. carotenemia
   D. hyperbilirubinemia
   E. markedly elevated CSF protein



6. Cranial CT is a valuable diagnostic tool in the evaluation of the following conditions **EXCEPT**

A. skull fractures
B. intracranial hemorrhages
C. acute infarcts
D. hydrocephalus
E. impending herniation

7. An approach to imaging of the spine in patients with cutaneous lesions is indicated in the following conditions **EXCEPT**

A. hairy patch
B. subcutaneous mass or lipoma
C. dermal sinus
D. coccygeal pits
E. scarlike lesions

8. Regarding myelomeningocele, all the following are true **EXCEPT**

A. risk of recurrence after one affected child is 3-4%
B. maternal periconceptional use of folic acid supplementation reduces the incidence of neural tube defects (NTDs) in pregnancies at risk by at least 50%
C. anticonvulsant valproic acid causes NTDs in approximately 1-2% of pregnancies when administered during pregnancy
D. hydrocephalus in association with a type II Chiari malformation develops in at least 80% of patients with myelomeningocele
E. the lower the deformity is in the neuraxis (sacrum), the more likely is the risk of hydrocephalus

9. Mobius syndrome is characterized by the following **EXCEPT**

A. bilateral facial weakness
B. paralysis of the abducens nerve
C. hypoplasia or agenesis of brainstem nuclei
D. feeding difficulties
E. mental retardation

10. Dandy-Walker malformation is characterized by the following **EXCEPT**

A. cystic dilatation of the fourth ventricle
B. herniation of the cerebellar tonsils though the foramen magnum
C. hypoplasia of the cerebellar vermis
D. hydrocephalus
E. an enlarged posterior fossa

11. Familial (autosomal recessive) microcephaly is characterized by the following **EXCEPT**

A. slanted forehead
B. prominent nose and ears
C. mild or borderline mental retardation
D. prominent seizures
E. surface convolutional markings of the brain

12. Causes of communicating hydrocephalus include

A. achondroplasia
B. aqueductal stenosis
C. Chiari malformation
D. Dandy-Walker malformation
E. Klippel-Feil syndrome

13. Nonobstructive or communicating hydrocephalus may follow the following conditions **EXCEPT**

A. a subarachnoid hemorrhage
B. pneumococcal meningitis
C. tuberculous meningitis
D. leukemic infiltrates
E. a vein of Galen malformation

14. Regarding the CSF, all the following are true **EXCEPT**

A. approximately 25% of CSF originates from extrachoroidal sources
B. in a normal child, about 20 mL/hr of CSF is produced
C. total volume of CSF approximates 150 mL in an infant
D. normally, CSF flows from the lateral ventricles through the foramina of Monro into the 3rd ventricle
E. CSF is absorbed primarily by the arachnoid villi

15. Trigoncephaly is a skull deformity that is a direct result of premature fusion of the following

A. sagittal suture
B. coronal suture
C. lambdoid suture
D. metopic suture
E. multiple sutures

16. Crouzon syndrome is characterized by the following **EXCEPT**

A. an autosomal dominant inheritance
B. brachycephaly

C. ocular proptosis
D. hypoplasia of the maxilla
E. syndactyly

17. Minor risk factors for recurrence of febrile seizure include the following **EXCEPT**
A. complex febrile seizure
B. age <1 yr
C. family history of febrile seizures
D. male gender
E. lower serum sodium

18. There are several predictors of epilepsy after febrile seizures, the highest percent of risk factor for subsequent epilepsy after febrile seizure is
A. complex febrile seizure, any type
B. fever <1 hr before febrile seizure
C. recurrent febrile seizures
D. focal complex febrile seizure
E. family history of epilepsy

19. Dravet syndrome is characterized by all the following **EXCEPT**
A. the most severe of the phenotypic spectrum of febrile seizures
B. its onset is in the 1st yr of life
C. seizures subsequently start to occur without fever
D. developmental delay
E. an autosomal dominant inheritance

20. Electroencephalogram (EEG) in febrile seizure is characterized by the following **EXCEPT**
A. an EEG need not norrmally be performed in first simple febrile seizure
B. an abnormal EEG could predict the future recurrence of febrile seizures or epilepsy
C. spikes during drowsiness are often seen in children with febrile seizures
D. an EEG performed within 2 wk of a febrile seizure often have nonspecific slowing
E. an EEG should be used to delineate the type of epilepsy

21. Absence seizures are **MOST** often initially treated with
A. ethosuximide
B. valproate
C. lamotrigine
D. acetazolamide
E. clonazepam

22. Benign myoclonic epilepsies are often best treated with
    A. clonazepam
    B. lamotrigine
    C. topiramate
    D. valproate
    E. benzodiazepines

23. Gingival hyperplasia, coarsening of the facies, hirsutism, and cerebellovestibular symptoms (nystagmus and ataxia) are adverse effects of
    A. carbamazepine
    B. lamotrigine
    C. phenytoin
    D. valproic acid
    E. succinimides

24. Rickets is a potential side effect from all the following **EXCEPT**
    A. phenytoin
    B. valproate
    C. phenobarbital
    D. primidone
    E. carbamazepine

25. Epilepsy surgery is often used to treat refractory epilepsy of a number of etiologies including the following **EXCEPT**
    A. cortical dysplasia
    B. tuberous sclerosis
    C. polymicrogyria
    D. degenerative problems
    E. Sturge-Weber syndrome

26. The **MOST** common cause of neonatal seizures is
    A. vascular events
    B. intracranial infections
    C. brain malformations
    D. hypoxic-Ischemic encephalopathy
    E. metabolic disturbances

27. All the following are true regarding diazepam in neonatal seizures **EXCEPT**
    A. diazepam is highly lipophilic
    B. it is cleared very quickly
    C. recurrence of seizures is more than other anticonvulsants
    D. it carries a risk of apnea and hypotension

E. it is currently recommended as a first-line agent

28. Neuro-imaging is warranted in a child with headache in the following conditions **EXCEPT**

A. abnormal neurologic examination
B. afternoon headache
C. headache in children <6 yr old
D. brief cough headache
E. migrainous headache in the child with no family history of migraine

29. Tension-type headaches (TTH) are characterized by the following **EXCEPT**

A. diffuse in location
B. not affected by activity
C. throbbing quality
D. mild to moderate in severity
E. less frequently associated with nausea and photophobia

30. The following are complications of neurofibromatosis (type 1) NF-1 **EXCEPT**

A. learning disability
B. cataracts
C. seizures
D. precocious puberty
E. scoliosis

31. Definite tuberous sclerosis complex (TSC) is diagnosed when at least 2 major or 1 major plus 2 minor features are present, all the following are minor features **EXCEPT**

A. shagreen patch
B. cerebral white matter migration lines
C. multiple dental pits
D. gingival fibromas
E. bone cysts

32. The facial port-wine stain of Sturge-Weber syndrome (SWS) is characterized by the following **EXCEPT**

A. present at birth
B. tends to be unilateral
C. always involves the upper face and eyelid
D. its distribution is consistent with the ophthalmic division of the trigeminal nerve
E. most children with facial port-wine stain have SWS

33. All the following are features of Von Hippel–Lindau (VHL) disease **EXCEPT**

A. its incidence is around 1: 36,000
B. fifty percent have a de novo gene mutation
C. hemangioblastoma of the spinal cord may be found
D. renal carcinoma is the most common cause of death
E. pheochromocytoma is a frequent association

34. PHACE syndrome denotes to the following **EXCEPT**
A. posterior fossa malformations
B. hemangiomas
C. anal anomalies
D. coarctation of the aorta
E. eye abnormalities

35. The current antiepileptic drug (AEDs) of choice for primary generalized tonic-clonic seizures in children is
A. phenytoin
B. carbamazepine
C. phenobarbital
D. valproate
E. topiramate

36. The **MOST** common type of cerebral palsy (CP) associated with seizures is
A. spastic hemiplegia
B. spastic quadriplegia
C. spastic diplegia
D. hypotonic CP
E. ataxic CP

37. The following are manifestations of increased intracranial pressure in an infant **EXCEPT**
A. increasing head circumference
B. bulging fontanel
C. failure to thrive
D. persistent vomiting
E. setting sun sign

38. Characteristic features of pseudotumor cerebri include the following **EXCEPT**
A. headache
B. stiff neck
C. papilledema
D. fatigue
E. abnormal CSF profile

39. How often are EEGs abnormal in healthy children?
A. 5%
B. 10%
C. 15%
D. 20%
E. 25%

40. The **MOST** common precipitant of status epilepticus in children is
A. CNS infection
B. fever
C. medication change
D. trauma
E. metabolic cause

41. A 9-month-old infant can do all the following **EXCEPT**
A. pulls to stand
B. pincer grasp
C. plays pat-a-cake
D. imitates sounds
E. comes when called

42. A premature infant blinks in response to a bright light at
A. 26 wk of corrected gestational age
B. 28 wk of corrected gestational age
C. 30 wk of corrected gestational age
D. 32 wk of corrected gestational age
E. 34 wk of corrected gestational age

43. The **MOST** common cause of anatomic megalencephaly is
A. Sotos syndrome
B. benign familial megalencephaly
C. Simpson-Golabi-Behmel syndrome
D. fragile X syndrome
E. Weaver syndrome

44. Craniosynostosis is defined as premature closure of the cranial sutures. Of the following, the **MOST** common form of craniosynostosis is
A. frontal plagiocephaly
B. occipital plagiocephaly
C. scaphocephaly
D. trigonocephaly
E. turricephaly

45. The following are risk factors for deformational plagiocephaly **EXCEPT**

A. female sex
B. firstborn child
C. congenital torticollis
D. developmental delay
E. exclusive bottle feeding

46. Major risk factors for recurrence of febrile seizure include

A. fever 38-39°c
B. family history of febrile seizures
C. family history of epilepsy
D. complex febrile seizure
E. lower serum sodium at time of presentation

47. In patients with febrile seizures, one of the following risk factors has the highest risk for subsequent epilepsy

A. recurrent febrile seizures
B. fever <1 hr before febrile seizure
C. neurodevelopmental abnormalities
D. family history of epilepsy
E. complex febrile seizures

48. The majority of patients who had prolonged febrile seizures and encephalopathy after vaccination and who had been presumed to have suffered from vaccine encephalopathy (seizures and psychomotor regression occurring after vaccination and presumed to be caused by it) turn out to have

A. generalized epilepsy with febrile seizures plus(GEFS+)
B. temporal lobe epilepsy secondary to mesial temporal sclerosis
C. myoclonic astatic seizures
D. Dravet syndrome
E. focal febrile seizures plus epilepsy variant

49. Higher risk of recurrence of the febrile seizure is associated with lower serum

A. sodium
B. potassium
C. chloride
D. calcium
E. magnesium

50. A history of personality change in a patient with seizure could suggest the following as a cause of seizure

A. intracranial tumor

B. degenerative disease
C. metabolic disease
D. stimulants drugs
E. congenital brain dysfunction

51. Guidelines on the evaluation of a first unprovoked nonfebrile seizure, the following studies are recommended in specific clinical situations
A. head CT
B. head MRI
C. spinal tap
D. ECG
E. EEG

52. Drug therapy should be based on the type of seizure and the epilepsy syndrome as well as on other individual factors.
Of the following, the drug of first choice for focal seizures and epilepsies is
A. carbamazepine
B. ethosuximide
C. valproate
D. lamotrigine
E. clobazam

53. Weight gain and alopecia are side effects of the following antiepileptic drugs AEDs
A. primidone
B. valproic acid
C. oxcarbazepine
D. lamotrigine
E. acetazolamide

54. The ketogenic diet is absolutely contraindicated in
A. primary carnitine deficiency
B. myoclonic– astatic epilepsy
C. tuberous sclerosis complex
D. Rett syndrome
E. infantile spasms

55. Discontinuation of antiepileptic drugs (AEDs) is usually indicated when children are free of seizures for at least
A. 1 yr
B. 1.5 yr
C. 2 yr
D. 2.5 yr

E. 3 yr

56. The following factors are associated with a higher risk of seizure relapse after antiepileptic drugs (AEDs) withdrawal **EXCEPT**

A. younger age of epilepsy onset
B. longer duration of epilepsy
C. presence of multiple seizure types
D. need to use more than 1 AED
E. abnormal EEG before medication is discontinued

57. Sudden unexpected death in epilepsy (SUDEP) is the most common epilepsy related mortality in patients with chronic epilepsy. All the following are risk factors **EXCEPT**

A. polypharmacology
B. female gender
C. age younger than 16 yr
D. long duration of epilepsy
E. frequent seizures

58. There are 5 main neonatal seizure types: subtle, clonic, tonic, spasms, and myoclonic.
One of the following seizures is frequently not associated with electrographic discharges

A. spasms
B. focal clonic
C. subtle
D. focal tonic
E. generalized myoclonic

59. Subtle seizures include all the following **EXCEPT**

A. blinking
B. mouthing
C. fluctuations in heart rate
D. hypotension episodes
E. apnea

60. The following are features of Aicardi syndrome **EXCEPT**

A. coloboma of the iris
B. retinal lacunae
C. agenesis of the corpus callosum
D. severe seizures
E. aminoaciduria

61. Many inborn errors of metabolism cause generalized convulsions in the newborn period. Prominent hiccups, persistent generalized seizures, and lethargy rapidly leading to coma are features of

A. propionic academia
B. maple syrup urine disease
C. nonketotic hyperglycinemia
D. Leigh disease
E. neonatal adrenoleukodystrophy

62. The initial drug used to control acute neonatal seizures is usually

A. diazepam
B. midazolam
C. phenobarbital
D. lorazepam
E. phenytoin

63. Intravenous phenytoin is not widely used to control acute neonatal seizures because of all the following **EXCEPT**

A. reduced solubility
B. severe local cutaneous reactions
C. interaction with other drugs
D. possible cardiac toxicity
E. not possible to mix with saline solutions

64. Status epilepticus is a medical emergency that should be anticipated in any patient who presents with an acute seizure. It is defined as continuous seizure activity or recurrent seizure activity without regaining of consciousness lasting for more than

A. 5 min
B. 15 min
C. 30 min
D. 45 min
E. 60 min

65. The following studies are needed for all patients with status epilepticus **EXCEPT**

A. glucose
B. magnesium
C. complete blood count
D. MRI
E. CT scan

66. Currently, the level of the evidence for refractory treatment of status epilepticus is strongest for

A. midazolam
B. propofol
C. levetiracetam
D. phenytoin
E. Phenobarbital

67. All the following are treatment options for breath-holding spells **EXCEPT**
A. education and reassurance of the parents
B. iron therapy
C. atropine sulfate
D. antiepileptic drug therapy
E. providing more care and interest for the patient

68. Migraine is the most frequent type of recurrent headache that is brought to the attention of parents and primary care providers, but it remains under recognized and undertreated, particularly in children.
Migraine is characterized by all the following **EXCEPT**
A. episodic attacks
B. moderate to severe in intensity
C. focal in location on the head
D. constant quality
E. may be associated with nausea and vomiting

69. The aura associated with migraine is a neurologic warning that a migraine is going to occur.
Of the following, the **LEAST** common type of typical auras is
A. sensory aura
B. dysphasic aura
C. visual aura
D. vertigo
E. distortion

70. The following are indications for neuroimaging in a child with a headache **EXCEPT**
A. abnormal neurologic examination
B. headache worst on first awakening
C. brief cough headache
D. frontal headache
E. migrainous headache in the child with no family history of migraine

71. Café-au-lait macules are not specific for NF-1; they may be seen in the following **EXCEPT**
A. Noonan syndrome

B. ataxia telangiectasia
C. Fanconi anemia
D. Gaucher disease
E. Hurler syndrome

72. The **MOST** frequent lesion associated with neurofibromatosis type 2 NF-2 is
A. bilateral vestibular schwannomas
B. intracranial
C. cataract
D. retinal hamartoma
E. skin plaque

73. Slow, writhing, continuous, and involuntary movements are called
A. stereotypies
B. tics
C. tremor
D. athetosis
E. chorea

74. A slowly progressive ataxia that involves the lower extremities to a greater degree than the upper extremities. The Romberg test result is positive; the deep-tendon reflexes are absent (particularly at the ankle), and the plantar response is typically extensor (Babinski sign).
Of the following, the **MOST** likely cause of this ataxia is
A. ataxia-telangiectasia
B. Friedreich ataxia
C. abetalipoproteinemia
D. Roussy-Levy disease
E. Ramsay Hunt syndrome

75. The **MOST** common acquired cause of chorea in childhood is
A. Huntington disease
B. paraneoplastic choreas
C. benign hereditary chorea
D. Sydenham chorea
E. chorea gravidarum

76. Prematurity is a major cause of
A. spastic diplegia
B. spastic quadriplegia
C. spastic hemiplegia
D. athetoid CP

E. dyskinetic CP

77. The **MOST** severe form of CP is
A. spastic diplegia
B. spastic quadriplegia
C. spastic hemiplegia
D. athetoid CP
E. dyskinetic CP

78. All the following can be used for treatment of cerebral palsy (CP) **EXCEPT**
A. benzodiazepines
B. baclofen
C. dantrolene
D. botulinum toxin
E. hyperbaric oxygen

79. Infants with Tay-Sachs disease (TSD) are characterized by the following **EXCEPT**
A. convulsions
B. blindness
C. deafness
D. cherry-red spots
E. microcephaly

80. Excessive irritability and crying, unexplained episodes of hyperpyrexia, vomiting, and difficulty feeding are the symptoms of the following neuro-degenerative disease
A. Tay-Sachs disease (TSD)
B. Sandhoff disease
C. juvenile GM2 gangliosidosis
D. Krabbe disease (KD)
E. metachromatic leukodystrophy

81. Presenting symptoms in pediatric multiple sclerosis (MS) include the following **EXCEPT**
A. hemiparesis
B. optic neuritis
C. ataxia
D. dysarthria
E. encephalopathy

82. The **MOST** common focal presentation of arterial ischemic stroke is
A. hemiparesis
B. acute visual deficit

C. speech deficit
D. sensory deficit
E. balance deficit

83. The **MOST** common cause of childhood subarachnoid and intraparenchymal hemorrhagic stroke (HS) is
A. moyamoya disease/syndrome
B. arteriovenous malformations
C. cerebral sinovenous thrombosis
D. hemolytic uremic syndrome
E. idiopathic thrombocytopenic purpura

84. Bilateral cortical visual dysfunction, encephalopathy, and seizures are seen in
A. global hypoxic–ischemic encephalopathy
B. herpes encephalitis
C. multiple sclerosis
D. hypertensive encephalopathy
E. hypoglycemia

85. The imaging studies are usually normal in the following stroke-like disorders of children
A. alternating hemiplegia
B. inborn errors of metabolism
C. hypertensive encephalopathy
D. hypoglycemia
E. global hypoxic–ischemic encephalopathy

86. Small vessel childhood primary angiitis of the CNS (SVcPACNS) is usually presented with
A. arterial ischemic stroke
B. seizures
C. cognitive dysfunction
D. personality changes
E. loss of social control

87. The diagnosis of diffuse CNS infections depends on examination of cerebrospinal fluid (CSF) obtained by lumbar puncture (LP). The cerebrospinal fluid (CSF) protein may reach 3,000 (mg/dL) in
A. acute bacterial meningitis
B. tuberculous meningitis
C. fungal meningitis
D. amebic (naegleria) meningoencephalitis

E. subdural empyema

88. The organism causing CNS infection is never seen on direct examination of CSF in
A. tuberculous meningitis
B. acute bacterial meningitis
C. partially treated bacterial meningitis
D. fungal meningitis
E. amebic (naegleria) meningoencephalitis

89. T-lymphocyte defects (congenital or acquired by chemotherapy, AIDS, or malignancy) are associated with an increased risk of infections of the CNS with
A. Listeria monocytogenes
B. Streptococcus pneumoniae
C. Neisseria meningitidis
D. Haemophilus influenzae type b
E. coagulase-negative Staphylococci

90. The CSF leukocyte count in normal healthy neonates may reach
A. 10 leukocytes/mm3
B. 15 leukocytes/mm3
C. 20 leukocytes/mm3
D. 25 leukocytes/mm3
E. 30 leukocytes/mm3

91. The following is an alternative treatment for L. monocytogenes meningitis
A. vancomycin
B. cefotaxime
C. chloramphenicol
D. intravenous trimethoprimsulfamethoxazole
E. meropenem

92. Reversible gallbladder pseudolithiasis, detectable by abdominal ultrasonography during treatment of bacterial meningitis is usually caused by
A. ceftazidime
B. cefotaxime
C. ceftriaxone
D. meropenem
E. chloramphenicol

93. Data support the use of intravenous dexamethasone, 0.15 mg/kg/dose given every 6 hr for 2 days, in the treatment of children older than 6 wk with acute bacterial meningitis caused by H. influenza type b. for the following reasons **EXCEPT**

A. shorter duration of fever
B. lower CSF protein
C. lower mortality
D. lower CSF lactate levels
E. reduction in sensorineural hearing loss

94. Seizures are common during the course of bacterial meningitis. Immediate therapy for seizures includes intravenous diazepam (0.1- 0.2 mg/kg/dose) or lorazepam (0.05-0.10 mg/kg/dose).
After immediate management of seizures, patients should receive the following anticonvulsant to reduce the likelihood of recurrence

A. valproic acid
B. phenytoin
C. phenobarbital
D. lorazepam
E. carbamazepine

95. The **MOST** common neurologic sequelae of bacterial meningitis is

A. recurrent seizures
B. delay in acquisition of language
C. hearing loss
D. cognitive impairment
E. visual impairment

96. The **MOST** common cause of viral meningoencephalitis are

A. arboviruses
B. enteroviruses
C. herpes simplex virus (HSV) type 1
D. varicella-zoster virus
E. mumps

97. The following viruses have clinical manifestations similar to that of the enteroviruses with the exception of more severe MRI lesions of the cerebral cortex and at times an absence of a CSF pleocytosis

A. arboviruses
B. parechoviruses
C. herpes simplex virus (HSV) type 1
D. rabies virus
E. Epstein-Barr virus

98. Detection of viral DNA or RNA by polymerase chain reaction is the test of choice in the diagnosis of CNS infection caused by

A. arboviruses
B. parechoviruses
C. West Nile virus (WNV)
D. rabies virus
E. Epstein-Barr virus

99. The predominant organisms causing brain abscesses in children are
A. aerobic and anaerobic streptococci
B. Streptococcus pneumoniae
C. Enterococcus faecalis
D. bacteroides spp
E. Haemophilus aphrophilus

100. A brain abscess can be treated with antibiotics without surgery in the following conditions **EXCEPT**
A. abscess is <2 cm in diameter
B. illness is of short duration (<2 wk)
C. lesion is located in the posterior fossa
D. no signs of increased intracranial pressure
E. child is neurologically intact

101. Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a clinical syndrome that mimics brain tumors and is characterized by the following **EXCEPT**
A. increased intracranial pressure ≥280 mm Hg in nonobese, nonsedated children
B. normal cerebrospinal fluid (CSF) cell count and protein content
C. normal to slightly decreased ventricular size
D. normal ventricular anatomy and position documented by MRI
E. papilledema

102. The following are hematologic causes of childhood pseudotumor cerebri **EXCEPT**
A. Wiskott-Aldrich syndrome
B. megaloblastic anemia
C. polycythemia
D. sickle cell disease
E. Fanconi anemia

103. The **MOST** common presenting complaint of intramedullary spinal cord tumors is
A. gait disturbance
B. sensory deficits
C. scoliosis
D. back pain

E. urinary urgency

104. The **MOST** common involved segments in transverse myelitis (TM) are in the

A. cervical region
B. thoracic region
C. lumbar region
D. lumbo-sacral region
E. sacral region

PART XXVII The Nervous System
ANSWERS
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1.**(D)**. In chronic subdural hemorrhages, the skull tends to assume a square or boxlike shape, because the long-standing presence of fluid in the subdural space causes enlargement of the middle fossa.
2.**(A)**. Although not a routine component of the examination, smell can be tested reliably as early as the 32nd wk of gestation by presenting a stimulus and observing for an alerting response, withdrawal, or both. Care should be taken to use appropriate stimuli, such as coffee or peppermint, as opposed to strongly aromatic substances (e.g., ammonia inhalants) that stimulate the trigeminal nerve. Each nostril should be tested individually by pinching shut the opposite side.
3.**(E)**. Thrombocytopenia with a platelet count $<20 \times 10^9$/L.
4.**(B)**. Normal CSF contains up to 5/mm^3 white blood cells (WBCs), and a newborn can have as many as 15/mm^3. Polymorphonuclear (PMN) cells are always abnormal in a child, but 1-2/mm^3 may be present in a normal neonate. An elevated PMN count suggests bacterial meningitis or the early phase of aseptic meningitis. CSF lymphocytosis can be seen in aseptic, tuberculous, or fungal meningitis; demyelinating diseases; brain or spinal cord tumor; immunologic disorders, including collagen vascular diseases; and chemical irritation (following myelogram, intrathecal methotrexate).
5.**(A)**. Normal CSF contains no red blood cells (RBCs); thus, their presence indicates a traumatic tap or a subarachnoid hemorrhage. Progressive clearing of the blood between the first and last samples indicates a traumatic tap. Bloody CSF should be centrifuged immediately. A clear supernatant is consistent with a bloody tap, whereas xanthochromia (yellow color that results from the degradation of hemoglobin) suggests a subarachnoid hemorrhage. Xanthochromia may be absent in bleeds <12 hr old, particularly when laboratories rely on visual inspection rather than spectroscopy. Xanthochromia can also occur in the setting of hyperbilirubinemia, carotenemia, and markedly elevated CSF protein.
6.**(C)**. CT is less useful for diagnosing acute infarcts in children, because radiographic changes might not be apparent for up to 24 hr.
7.**(D)**. Imaging of the spine is not indicated in patients with simple dimples (<5 mm, <25 mm from anal verge) or coccygeal pits.
8.**(E)**. Generally, the lower the deformity is in the neuraxis (sacrum), the less likely is the risk of hydrocephalus.
9.**(E)**. The immobile, dull facies might give the incorrect impression of mental retardation; the prognosis for normal development is excellent in most cases.

10.**(B)**. Chiari malformation is the most common malformation of the posterior fossa and hindbrain. It consists of herniation of the cerebellar tonsils though the foramen magnum.
11.**(C)**. Familial (autosomal recessive) microcephaly is characterized by severe mental retardation.
12.**(A)**. Aqueductal stenosis, Chiari malformation, Dandy-Walker malformation, and Klippel-Feil syndrome are causes of non-communicating hydrocephalus.
13.**(E)**. A vein of Galen malformation can expand to become large and, because of its midline position, obstruct the flow of CSF.
14.**(C)**. The total volume of CSF approximates 50 mL in an infant and 150 mL in an adult.
15.**(D)**.
Sagittal= Dolicocephaly or scaphocephaly (boat-shaped)
Coronal= Unilateral: plagiocephaly Bilateral: brachycephaly, acrocephaly
Lambdoid= Lambdoid/occipital plagiocephaly; right side affected in 70% of cases
Multiple= Oxycephaly
16.**(E)**. Apert syndrome is characterized by syndactyly of the 2nd, 3rd, and 4th fingers, which may be joined to the thumb and the 5th finger.
17.**(B)**. Age <1 yr, duration of fever <24 hr, and fever 38-39^{0}C are major risk factors for recurrence of febrile seizure.
18.**(D)**. Focal complex febrile seizure has 29% risk for subsequent epilepsy.
19.**(E)**. This syndrome is usually caused by a new mutation, although rarely it is inherited in an autosomal dominant manner. The mutated gene is located on 2q24-31 and encodes for SCN1A, the same gene mutated in GEFS+ spectrum. However, in Dravet syndrome the mutations lead to loss of function and thus to a more severe phenotype.
20.**(B)**. An EEG would not predict the future recurrence of febrile seizures or epilepsy even if the result is abnormal.
21.**(A)**. Absence seizures are most often initially treated with ethosuximide, which is as effective as and less toxic than valproate and more effective than lamotrigine.
22.**(D)**. Benign myoclonic epilepsies are often best treated with valproate, particularly when patients have associated generalized tonic-clonic and absence seizures. Benzodiazepines, clonazepam, lamotrigine, and topiramate are alternatives for the treatment of benign myoclonic epilepsy.
23.**(C).**
24.**(B)**. Potential side effects are rickets from phenytoin, phenobarbital, primidone, and carbamazepine (enzyme inducers that reduce 25-hyrdroxy-vitamin D level by inducing its metabolism) and hyperammonemia from valproate.
25.**(D)**. Epilepsy surgery is often used to treat refractory epilepsy of a number of etiologies including cortical dysplasia, tuberous sclerosis, polymicrogyria, hypothalamic hamartoma, and hemispheric syndromes, such as Sturge-Weber syndrome, hemimegalencephaly, Rasmussen encephalitis, and Landau-Kleffner syndrome. Patients

with intractable epilepsy resulting from metabolic or degenerative problems are not candidates for resective epilepsy surgery.
26.**(D)**. Hypoxic-ischemic encephalopathy is the most common cause of neonatal seizures, accounting for 50-60% of patients. Seizures secondary to this encephalopathy occur within 12 hr of birth.
27.**(E).** Because of the respiratory and blood pressure limitations and because the intravenous preparation contains sodium benzoate and benzoic acid, it is currently not recommended as a first-line agent.
28.**(B)**. Neuroimaging is warranted when the neurologic examination is abnormal or unusual neurologic features occur during the migraine; when the child has headaches that awaken him or her from sleep or that are present on first awakening and remit with upright posture; when the child has brief headaches that only occur with cough or bending over; and when the child has migrainous headache with an absolutely negative family history of migraine or its equivalent (e.g., motion sickness, cyclic vomiting).
29.**(C)**. TTH are mild to moderate in severity, are diffuse in location, are not affected by activity (although the patient may not feel like being active), and are nonthrobbing (often described as a constant pressure).
30.**(B)**. Cataracts are found in 60-81% of patients with NF-2.
31.**(A)**. Shagreen patch is a major feature.
32.**(E)**. It is important to note that not all children with facial port-wine stain have SWS. In fact, the overall incidence of SWS has been reported to be 8-33% in those with a port-wine stain.
33.**(B)**. Around 80% of individuals with VHL disease have an affected parent, and around 20% have a de novo gene mutation.
34.**(C).** Arterial anomalies.
35.**(D)**. Class I evidence demonstrates that phenytoin, carbamazepine, phenobarbital, primidone, valproate, topiramate, oxcarbazepine, and lamotrigine are effective for the treatment of primary generalized tonic-clonic seizures. On the basis of efficacy and tolerability, valproate and lamotrigine have emerged as the current drugs of choice for grand mal seizures.
36.**(A).**
37.**(D)**. The manifestations of increased intracranial pressure in an infant as compared with an older child are:
Infant**:** Increasing head circumference, delayed closure of the fontanel, suture separation, bulging fontanel, failure to thrive, macrocephaly, setting sun sign, and shrill cry.
Older child**:** Headache (especially in the early morning, awakening the child from sleep, or association with vomiting), nausea, persistent vomiting, personality/mood changes, lethargy, anorexia, fatigue, somnolence, diplopia as a result of sixth-nerve palsy or third-nerve palsy with uncal herniation, and papilledema.
38.**(E)**. Normal CSF profile with the exception of an elevated opening pressure.

39.**(B)**. Approximately 10% of "normal" children have mild, nonspecific abnormalities in background activity.
40.**(B)**. Fever/infection (36%) ,CNS infection (5%) , medication change (20%) , trauma (4%) , unknown (9%) , cerebrovascular (3%) , metabolic (8%) , ethanol/drug-related (2%) , congenital (7%) , tumor (1%) , and anoxia (5%).
41.**(E)**. Walks with 1 hand held, releases an object on command, comes when called, and 1-2 meaningful words are milestones of 12-month-old child.
42.**(B)**. At 28 wk of corrected gestational age, a premature infant blinks in response to a bright light, and at 32 wk, the infant maintains eye closure until the light source is removed. A normal 37 wk infant turns the head and eyes toward a soft light, and a term infant is able to fix on and follow a target, such as the examiner's face.
43.**(B)**.
44.**(C)**. Premature closure of the sagittal suture produces a long and narrow skull, or scaphocephaly**,** the most common form of craniosynostosis.
45.**(A)**. Male sex.
46.**(A)**. Risk factors for recurrence of febrile seizure include
MAJOR

- Age <1 yr
- Duration of fever <24 hr
- Fever 38-39°C (100.4-102.2°F)

MINOR

- Family history of febrile seizures
- Family history of epilepsy
- Complex febrile seizure
- Daycare
- Male gender
- Lower serum sodium at time of presentation

47. **(C)**. Risk factor for subsequent epilepsy
* simple febrile seizure 1%
* recurrent febrile seizures 4%
* complex febrile seizures (more than 15 min duration or recurrent within 24 hr) 6%
* fever<1 hr before febrile seizure 11%
* family history of epilepsy 18%
* complex febrile seizures (focal) 29%
* neurodevelopmental abnormalities 33%

48.**(D)**. The majority of patients who had prolonged febrile seizures and encephalopathy after vaccination and who had been presumed to have suffered from vaccine encephalopathy(seizures and psychomotor regression occurring after vaccination and presumed to be caused by it) turn out to have Dravet syndrome mutations, indicating that their disease is caused by the mutation and not secondary to

the vaccine. This has raised doubts about the very existence of the entity termed vaccine encephalopathy.

49.**(A)**. A low sodium level is associated with higher risk of recurrence of the febrile seizure within the following 24 hr.

50.**(A)**. A history of personality change or symptoms of increased intracranial pressure can suggest an intracranial tumor. Similarly, a history of cognitive regression can suggest a degenerative or metabolic disease. Certain medications such as stimulants or antihistamines, particularly sedating ones, can precipitate seizures. A history of prenatal or perinatal distress or of developmental delay can suggest etiologic congenital or perinatal brain dysfunction.

51.**(C).** Spinal tap is considered in patients with suspected meningitis or encephalitis, in children without brain swelling or papilledema, and in children in whom a history of intracranial bleeding is suspected without evidence of such on head CT.

52.**(A)**. In general, the drugs of first choice for focal seizures and epilepsies are oxcarbazepine and carbamazepine; for absence seizures, ethosuximide; for juvenile myoclonic epilepsy, valproate and lamotrigine; for Lennox-Gastaut syndrome, clobazam, valproate, topiramate, lamotrigine, and, most recently, as add on, rufinamide; and for infantile spasms, adrenocorticotropic hormone (ACTH).

53.**(B)**. Side effects of valproic acid: weight gain, hyperammonemia, tremor, alopecia, and menstrual irregularities. Serious: hepatic and pancreatic toxicity.

54.**(A)**. The ketogenic diet is absolutely contraindicated in carnitine deficiency (primary); carnitine palmitoyltransferase I or II deficiency; carnitine translocase deficiency; β-oxidation defects; medium-chain acyl dehydrogenase deficiency; long-chain acyl dehydrogenase deficiency; short-chain acyl dehydrogenase deficiency; long-chain 3-hydroxyacyl-coenzyme A deficiency; medium-chain 3-hydroxyacyl-coenzyme A deficiency; pyruvate carboxylase deficiency; and porphyrias.

55.**(C)**. Discontinuation of AEDs is usually indicated when children are free of seizures for at least 2 yr.

56.**(A)**. Older age of epilepsy onset.

57.**(B)**. Male gender.

58.**(C)**. Spasms, focal clonic, focal tonic, and generalized myoclonic seizures are, as a rule, associated with electrographic discharges (epileptic seizures), whereas motor automatisms, the subtle, generalized tonic and multifocal myoclonic episodes are frequently not associated with discharges and thus are thought to often represent release phenomena with abnormal movements secondary to brain injury rather than true epileptic seizures.

59.**(D)**. Subtle seizures include transient eye deviations, nystagmus, blinking, mouthing, abnormal extremity movements (rowing, swimming, bicycling, pedaling, and stepping), and fluctuations in heart rate, hypertension episodes, and apnea. Subtle seizures occur more commonly in premature than in full-term infants.

60.**(E)**. Brain malformations account for 5-10% of neonatal seizure cases. An example is Aicardi syndrome, which affects girls only and consists of retinal lacunae, agenesis of

the corpus callosum, and severe seizures including subsequent infantile spasms with hypsarrhythmia that is sometimes initially unilateral on EEG.
61.**(C)**. Nonketotic hyperglycinemia, an intractable condition characterized by markedly elevated plasma and CSF glycine levels, prominent hiccups, persistent generalized seizures, and lethargy rapidly leading to coma.
62.**(D)**. The initial drug used to control acute seizures is usually lorazepam. Lorazepam is distributed to the brain very quickly and exerts its anticonvulsant effect in <5 min. It is not very lipophilic and does not clear out from the brain very rapidly. Its action can last 6-24 hr. Usually, it does not cause hypotension or respiratory depression. The dose is 0.05 mg/kg (range: 0.02-0.10 mg/kg) every 4-8 hr.
63.**(E)**. It is not possible to mix phenytoin or fosphenytoin with dextrose solutions.
64.**(A)**.
65.**(D)**. Lumbar puncture, comprehensive toxicologic screens, MRI, and other laboratory tests are performed depending on clinical suspicion and need.
66.**(A)**. Currently, the level of the evidence for refractory treatment is strongest for midazolam and valproate, followed by propofol and pentobarbital/thiopental, followed by levetiracetam, phenytoin/fosphenytoin, lacosamide, topiramate, and phenobarbital.
67.**(E)**. All parents should be taught not to provide secondary gain when the episodes occur, because this can reinforce the episodes.
68.**(D)**. Migraine is characterized by episodic attacks that may be moderate to severe in intensity, focal in location on the head, have a throbbing quality, and may be associated with nausea, vomiting, light sensitivity, and sound sensitivity. Compared to adults, pediatric migraine is shorter in duration and has a bilateral, often bifrontal, location. Migraine can also be associated with an aura that may be typical (visual, sensory, or dysphasic) or atypical (i.e., hemiplegic, “Alice in Wonderland” syndrome).
69.**(B)**. Dysphasic auras are the least-common type of typical aura and have been described as an inability or difficulty to respond verbally.
70.**(D)**. When the headache is mostly in the occipital area.
71.**(E)**. Hunter syndrome.
72.**(A)**. Bilateral vestibular schwannomas 90-95%.
73.**(D)**.
74.**(B)**. Friedreich ataxia is inherited as an autosomal-recessive disorder involving the spinocerebellar tracts, dorsal columns in the spinal cord, the pyramidal tracts, and the cerebellum and medulla. The onset of ataxia is somewhat later than in ataxiatelangiectasia, but usually occurs before age 10 yr. The ataxia is slowly progressive and involves the lower extremities to a greater degree than the upper extremities. The Romberg test result is positive; the deep-tendon reflexes are absent (particularly at the ankle), and the plantar response is typically extensor (Babinski sign). Patients develop a characteristic explosive, dysarthric speech, and nystagmus is present in most children.

75.**(D)**. Sydenham chorea (St. Vitus dance) is the most common acquired chorea of childhood. It occurs in 10-20% of patients with acute rheumatic fever, typically weeks to months after a group A β-hemolytic streptococcal infection. Peak incidence is at age 8-9 yr, with a female predominance of 2:1.
76.**(A)**. Prematurity, ischemia, infection, and endocrine/metabolic (e.g., thyroid) are major causes of spastic diplasia.
77.**(B)**. Spastic quadriplegia is the most severe form of CP because of marked motor impairment of all extremities and the high association with intellectual disability and seizures.
78.**(E)**. Hyperbaric oxygen has not been shown to improve the condition of children with CP.
79.**(E)**. Macrocephaly becomes apparent by 1 yr of age and results from the 200-300 fold normal content of GM2 ganglioside deposited in the brain.
80.**(D)**. The symptoms of KD become evident in the 1st few mo of life and include excessive irritability and crying, unexplained episodes of hyperpyrexia, vomiting, and difficulty feeding. In the initial stage of KD, children are often treated for colic or milk allergy with frequent formula changes.
81.**(E)**. Encephalopathy is less common and suggests consideration of ADEM or possibly neuromyelitis optica (NMO).
82.**(A)**. The most common focal presentation is hemiparesis, but acute visual, speech, sensory, or balance deficits also occur. Children with these presentations require urgent neuroimaging and consultation with a child neurologist, as emergency interventions may be indicated.
83.**(B)**. Arteriovenous malformations are the most common cause of childhood subarachnoid and intraparenchymal HS and may occur anywhere.
84.**(D)**. The posterior reversible leukoencephalopathy syndrome is seen in children with hypertension, often in the context of an acute rise in blood pressure. Posterior regions are selectively involved, possibly resulting in symptoms of bilateral cortical visual dysfunction in addition to encephalopathy and seizures.
85.**(A)**.
86.**(B)**. Seizures are a hallmark of SVcPACNS, as more than 80% of children with SVcPACNS present with seizures; all seizure types are seen.
87.**(B)**. In tuberculous meningitis, CSF protein is 100-3,000; may be higher in presence of block, while in other options, it may reach 500.
88.**(A)**. Acid-fast organisms almost never seen on smear. Organisms may be recovered in culture of large volumes of CSF. Mycobacterium tuberculosis may be detected by PCR of CSF.
89.**(A).**
90.**(E)**. Normal healthy neonates may have as many as 30 leukocytes/mm3 (usually <10), but older children without viral or bacterial meningitis have <5 leukocytes/mm3 in the CSF; in both age groups there is a predominance of lymphocytes or monocytes.

91.**(D)**. If L. monocytogenes infection is suspected, as in young infants or those with a T-lymphocyte deficiency, ampicillin (200 mg/kg/24 hr, given every 6 hr) should also be given because cephalosporins are inactive against L. monocytogenes. Intravenous trimethoprim sulfamethoxazole is an alternative treatment for L. monocytogenes.
92.**(C)**. Ceftriaxone may cause reversible gallbladder pseudolithiasis, detectable by abdominal ultrasonography. This is usually asymptomatic but may be associated with emesis and upper right quadrant pain.
93.**(C)**. Among children with meningitis caused by H. influenza type b, corticosteroid recipients have a shorter duration of fever, lower CSF protein and lactate levels, and a reduction in sensorineural hearing loss. Data in children regarding benefits, if any, of corticosteroids in the treatment of meningitis caused by other bacteria are inconclusive.
94.**(B)**. After immediate management of seizures, patients should receive phenytoin (15-20 mg/ kg loading dose, 5 mg/kg/24 hr maintenance) to reduce the likelihood of recurrence. Phenytoin is preferred to phenobarbital because it produces less CNS depression and permits assessment of a patient's level of consciousness. Serum phenytoin levels should be monitored to maintain them in the therapeutic range (10-20 µg/mL).
95.**(C)**. Sensorineural hearing loss is the most common sequela of bacterial meningitis and, usually, is already present at the time of initial presentation. It is a result of cochlear infection and occurs in as many as 30% of patients with pneumococcal meningitis, 10% with meningococcal, and 5-20% of those with H. influenzae type b meningitis. Hearing loss may also be caused by direct inflammation of the auditory nerve. All patients with bacterial meningitis should undergo careful audiologic assessment before or soon after discharge from the hospital. Frequent reassessment on an outpatient basis is indicated for patients who have a hearing deficit.
96.**(B)**. Enteroviruses are the most common cause of viral meningoencephalitis. As of 2014, more than 70 serotypes of these small RNA viruses have been identified.
97.**(B)**. Parechoviruses may be an important cause of aseptic meningitis or encephalitis in infants. The clinical manifestations are similar to that of the enteroviruses with the exception of more severe MRI lesions of the cerebral cortex and at times an absence of a CSF pleocytosis.
98.**(B)**. Detection of viral DNA or RNA by polymerase chain reaction is the test of choice in the diagnosis of CNS infection caused by HSV, parechovirus, and enteroviruses, respectively. CSF serology is the diagnostic test of choice for WNV.
99.**(A)**. The predominant organisms causing brain abscesses in children are aerobic and anaerobic streptococci (60-70% of the cases) with Streptococcus milleri gp (Streptococcus anginosus, Streptococcus constellatus, and Streptococcus intermedius) being increasingly isolated from surgically drained brain abscesses.
100.**(C).** Surgery is indicated when the abscess is >2.5 cm in diameter, gas is present in the abscess, the lesion is multiloculated, the lesion is located in the posterior fossa, or a fungus is identified. Associated infectious processes, such as mastoiditis, sinusitis, or a

periorbital abscess, may require surgical drainage. The duration of antibiotic therapy depends on the organism and response to treatment but is usually 4-6 wk.

101.**(A)**. Increased intracranial pressure ≥280 mm Hg in sedatedor obese children; ≥250 mm Hg in nonobese, nonsedated children.

102.**(B)**. Iron-deficiency anemia.

103.**(D)**. With the exception of the uncommon malignant glial tumors of the spinal cord, which tend to present precipitously, intramedullary spinal cord tumors present in a very insidious manner. Back pain related to the level of the tumor is a common presenting complaint. It is likely that this pain will awaken the child from sleep and improve as the day progresses.

104.**(B)**.